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Case Report

Extra salivary Adenoid Cystic Carcinoma; report of two cases

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INTRODUCTION

Adenoid cystic carcinoma is a well-recognized malignant neoplasm of both the major and minor salivary glands. It carries a poor prognosis, and may also occur as a primary neoplasm elsewhere. Adenoid cystic carcinoma in the breast is uncommon, accounting for 0.1% of all breast cancers.[1-3] It has very favorable biological characteristics and has an excellent prognosis. Tumour recurrences as well as regional and distant metastases have been described rarely. The first patient in our report had a very short history of a breast lump and radiologically confirmed bilateral pulmonary metastases even though she had a node negative, histologically proven low grade tumour.

Primary adenoid carcinoma of the skin is extremely rare. It may present as a primary site or as distant metastasis. They are locally aggressive mimicking their salivary counterpart with a recurrence rate of 51%.[4] Our second case is a patient with an eight year history of cutaneous swelling diagnosed clinically as a sebaceous cyst and confirmed as adenoid cystic carcinoma on pathology.

CASE REPORTS

CASE 1 A 73 year old woman was referred, complaining of mastodynia and a lump in the left breast for four months. Examination revealed an irregular, hard, tender mass behind and lateral to the left nipple, measuring 4 x 4 cm in size. No axillary lymphadenopathy was detected on examination. Mammogram showed a 3.5 cm ill defined lobulated mass above and lateral to the left nipple. Appearances were those of a carcinoma. Fine needle aspiration cytology (*fig 1*) showed an aspirate of high cellularity with groups of monomorphic cells associated with round to oval globules of matrix material. In the background were scattered single cells. The appearance was highly suggestive of adenoid cystic carcinoma. She had a core biopsy, which confirmed an adenoid cystic carcinoma.

Preoperative CXR revealed multiple lung deposits. Subsequent CT scan of chest and abdomen showed bilateral multiple lung nodules suggestive of metastases (*fig 2*). The patient underwent left a total mastectomy and axillary node clearance. Histopathology was consistent with the preoperative diagnosis of adenoid cystic carcinoma. Of the 22 axillary nodes removed none contained metastatic deposits. The tumour was negative for oestrogen and progesterone receptors and no lymphovascular invasion was seen. She made a satisfactory postoperative recovery. Neither chemotherapy nor hormonal therapy was required after consultation with oncology. At six months of follow up she is well with no recurrence.

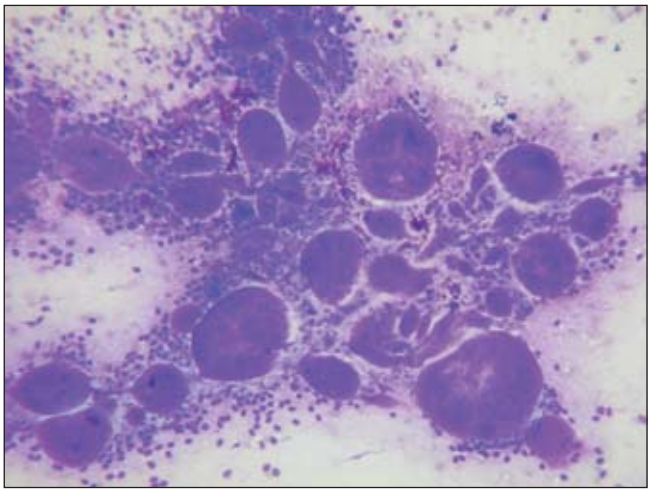

Fig 1. FNA cytological appearance of adenoid cystic carcinoma with globules of matrix material surrounded by monomorphic epithelial cells. MGG stain, x 200 mag.

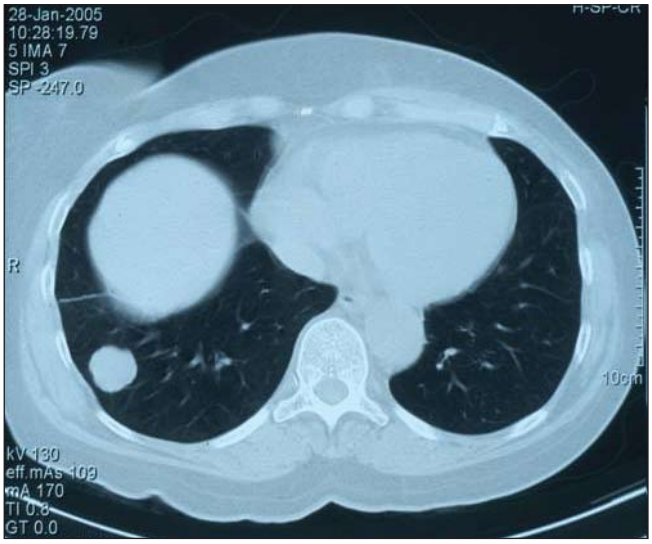

Fig 2. CT Scan of chest showing one of the lung nodules.

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CASE 2 A 47 year old woman was referred with a small painless slowly growing swelling on her right upper back for eight years. This was clinically diagnosed as a sebaceous cyst. This lesion was excised under local anesthesia. Histology (*fig 3*) of the excised lesion revealed adenoid cystic carcinoma.

The tumour consisted of a cribriform proliferation of small dark basaloid cells with hyaline membrane type material in some of the cribriform proliferations. The lesion had a lobulated architecture and infiltrative margins. The periphery of the lesion showed perineural invasion. Immunohistochemical stains showed co-expression of smooth muscle actin, S100, and CK5 & 6 within the myoepithelial component of the adenoid cystic carcinoma. The lesion was re-excised with wider margins. She recovered satisfactorily and eight months after surgery there were no signs of recurrence.

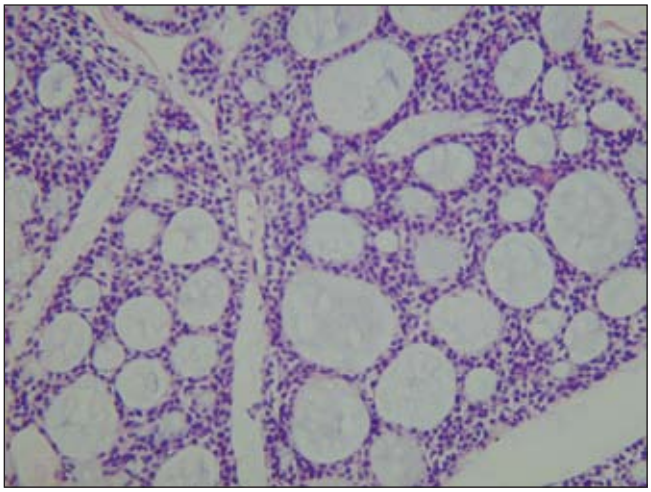

Fig 3. Histology of adenoid cystic carcinoma, cribriform growth pattern. H&E x 200 mag.

DISCUSSION

Adenoid cystic carcinoma is a rare neoplasm of the breast. It occurs predominantly in women aged 50-60 years and may be bilateral.[5] It tends to develop in the periareolar area. Patients may present with a slow growing solid mass for months or years without distant spread. Another common feature is intermittent pain and tenderness in the breast mass. Mammogram and ultrasonography findings vary widely and are not diagnostic.[3] These tumours often appear as a small lobulated nodule with clearly defined margins.[6, 7] They may also show as large masses with more ill defined margins. In this case mammogram showed an ill defined, lobulated lesion.

Cytology and histological appearances of adenoid cystic carcinoma in the breast are similar to the adenoid cystic carcinoma of other anatomical sites. Recent reports described characteristic features on FNA cytology to enable preoperative diagnosis.[1, 2, 8, 9] Cellular aspirates with tightly cohesive aggregates of cells with enclosed spheres and interconnecting cylinders of acellular material are characteristic. The principal cell type represent the epithelial cells and a minor proportion of cells are ovoid to spindle shaped with hyperchromatic nuclei, representing myoepithelial cells. Another characteristic feature is the numerous bare nuclei in the background. Two distinctive histological features of adenoid cystic carcinoma of the breast are the intercellular cystic spaces lined by basement membrane material and biphasic cellularity with myoepithelial cells intermixed with another cell type.[3] Immunohistochemical staining confirms the presence of a dual population of epithelial and myoepithelial cells.[1, 8, 10]

Previous reports have stressed the excellent prognosis for patients with adenoid cystic carcinoma of breast; however tumour recurrence and distant metastases have been described on rare occasions.[4] Some reports show clear cut evidence of malignancy with documented potential for metastases, others show benign behaviour and the less well defined groups reside between these two extremes. Qizilbash[11] reviewed 95 well documented cases with only one case of lymph node metastases documented. Six cases of distant metastases are recorded, (five had pulmonary metastases).

Several studies have investigated the possible correlation between histological grade and prognosis. Some reports concluded that a solid variant of mammary adenoid cystic carcinoma had a more aggressive clinical course. Leeming[12] reviewed 123 cases in the literature and noted that several features distinguished adenoid cystic carcinoma from other breast tumours. Prognosis appeared to be favourable and axillary node involvement was uncommon. Distant metastases were unusual and tended to occur without previous lymph node involvement. Tumours rarely showed positivity for oestrogen and progesterone receptors. There was relative absence of the perineural invasion which characterizes lesions in the salivary glands.

There is no consensus on optimal treatment for adenoid cystic carcinoma of the breast. Surgical management has evolved from radical mastectomy to breast conserving surgery. Due to the documented recurrence of the tumour after local excision, simple mastectomy with careful follow up is recommended. The role of radiotherapy, chemotherapy or hormonal therapy is unproven.

The second patient in our report had an asymptomatic swelling on her back for eight years. Histology was consistent with adenoid cystic carcinoma. Multiple treatment modalities including surgery, radiation and chemotherapy should be used for locally aggressive and potentially metastasizing adenoid cystic carcinoma of the skin. This case demonstrates the benefit of sending even benign lesions for histology.

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